

Cardiac Anomalies

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A CLINICOPATHOLOGIC CORRELATION

by

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Preface

Congenital cardiac malformations have been in the medical limelight for the past few years. The reason for this special interest has been the great success in the surgical treatment of some of these anomalies. Moreover increasing numbers of cardiac defects are becoming operable as new surgical techniques are being developed.

In order to give the individual patient the best that surgery can offer it is necessary that an accurate diagnosis be made in each instance. To accomplish this however physical findings must be correlated with the radiologic, physiologic and electrocardiographic findings.

Since the problem is fundamentally one of abnormal morphology of the heart we have based the classification and the entire correlation within each entity on the features shown by the actual cardiac specimens. We have used exclusively material from cases verified either at surgery or at post mortem examination.

Each anomaly has been covered with brief statements regarding the pathology, course of the circulation, clinical features, laboratory findings, electrocardiography, radiologic studies, prognosis as well as the medical and surgical treatment. Thus we hope this book will be of value not only to the medical student but to the pediatrician and general practitioner as well.

A unique feature of the book is the presentation of microphotographs of the lungs and great vessels. The changes produced in these structures by certain cardiac anomalies are frequently severe enough to influence the prognosis and the surgical management.

We wish to express our thanks to the physicians of St. Louis and the State of Missouri for sending us many interesting cases as well as many intriguing autopsy specimens. Our appreciation is also extended to the Department of Radiology of the St. Mary's Group of Hospitals of St. Louis University for its excellent cooperation. The assistance of Dr. N. Staples, T. D. Brewer, A. Freitag and Mrs. James Donahoe in preparing the illustrations is sincerely appreciated.

VINCENT MORACUES
CHESTER P. LINXWILER

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Figures 22, 23, 24, 25 from V Moragues, "Persistent Common Atrioventricular Ostium," American Heart Journal, Vol 25, No 1, pages 123-127, January, 1943

Figures 34, 35, 36, 37, 38, from Ebel, Lynxwiler and Moragues, "Tricuspid Atresia with Rudimentary Right Ventricle," Journal of Pediatrics, Vol 39, No 2, pages 211-215, August, 1951

Figures 41, 42 from Inkley, Moragues and Lynxwiler, "Tricuspid Atresia," St Louis University Bulletin Vol 2, No 6, June, 1950

Figures 65, 66, 67, 68, 69, 70, 71, 72, 73 from V Moragues, "Persistent Truncus Arteriosus," American Journal of Clinical Pathology, Vol 20, No 9, September, 1950

Figures 130, 131 from Lynxwiler, Smith and Babich, "Coarctation of the Aorta," Archives of Pediatrics, Vol 68 No 5, pages 203-207, May, 1951

Figures 132, 133, 134 from Lynxwiler, Smith, Lucido and Inkley, "Coarctation of the Aorta," Journal of the Missouri State Medical Association, pages 177-180 March, 1951

Figures 139, 140, 141 from Moragues Moore and Rossen, "Coarctation of Aorta," American Heart Journal, Vol 24, No 6, pages 828-834, December, 1942

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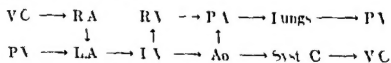
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Introduction

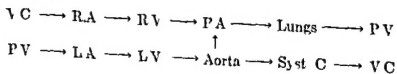
During fetal life the foramen ovale and the ductus arteriosus provide shunting channels through which the pulmonary circulation is by-passed to a great extent. In reality the fetal circulation can be considered as consisting of a single circuit. It is no wonder therefore that even the most formidable malformations of the heart are seldom a cause of intrauterine death. At birth however with the ligation of the umbilical cord and the expansion of the lung, the circulation changes from a single circuit to a double circuit in parallel (systemic and pulmonary). This change entails enormous hemodynamic readjustments which are not possible in the more severely deformed hearts. *This is the reason why the greatest mortality from congenital heart disease occurs in the neonatal period.*

In general there are three mechanisms of adaptation by which the circulation adjusts itself to the altered morphology present in the heart. The *first* of these mechanisms is *complete pooling* of the arterial and venous blood. This mechanism operates in such malformations as cor triloculare, cor biloculare, tricuspid atresia, pulmonary atresia and atresia of the aortic or mitral valve. Complete pooling as a mechanism of adaptation is understood when one considers the course of the circulation in tricuspid valve atresia.



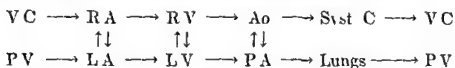
As the tricuspid valve is atretic, all the blood entering the right auricle must pass to the left auricle either through an auricular septal defect or a patent foramen ovale. Therefore there is complete pooling of arterial and venous blood in the left side of the heart.

The second mechanism of adaptation is that of a *unidirectional* shunt which occurs in cases of persistent ductus arteriosus, isolated septal defects and in cases in which the pulmonary artery or aorta 'over-ride' a ventricular septal defect. In such instances the unidirectional shunts are well tolerated as the shunted blood returns to the side of the circulation from which it started. Unidirectional shunts are easily understood when one considers the course of the circulation in a case of patent ductus arteriosus.



Since the pressure in the aorta exceeds the pressure in the pulmonary artery, the flow through the patent ductus is from the systemic to the pulmonary circulation. The blood shunted through the ductus returns to the systemic (left) side of the heart through the pulmonary veins, hence, hemodynamic equilibrium is maintained.

Multidirectional shunts represent the *third* mechanism by which circulatory adaptation occurs. These multiple shunts are necessary in cases of complete transposition of the arterial trunks, since in these conditions the shunted blood fails to return to its original side of the circulation. To illustrate this mechanism, let us consider the course of the circulation in a case of complete transposition of the arterial trunks with associated septal defects and a patent ductus arteriosus.



Since in this condition the two circulations exist as entirely separate circuits, life can be maintained only by the shunting of sufficient oxygenated blood into the systemic circulation. However, since the shunted blood does not return to the side of the circulation from which it originated, reverse shunts are necessary to maintain blood volume equilibrium in the two circulations. It is well known that patients with complete transposition of the arterial trunks live longer when more than one shunt is present.

In our opinion, the above mechanisms of circulatory adaptation are of such importance in the understanding of the hemodynamics in each individual case that they will be referred to again in the various chapters of this book.

Most malformations of the heart result in alterations in the pulmonary circulation which interfere with the proper oxygenation of the blood. Some of the conditions result in a decreased flow and a decreased pressure in the pulmonary circuit with a consequent oxygen unsaturation of the arterial blood from birth (congenital cyanosis). Examples: Tetralogy of Fallot, Pulmonary valvular atresia, Tricuspid atresia, etc. Other malformations result in an increased flow and an increased pressure in the pulmonary circulation with subsequent development of pulmonary arteriosclerosis (delayed cyanosis). Examples: occasional Patent ductus arteriosus, Eisenmenger's complex, etc.

To give objective evidence of the changes in the pulmonary circulation in these conditions, a number of lung sections and sections of the aorta and the pulmonary artery have been included in this atlas.

1

Malpositions of the Heart

Pathology

Two types

- a) Isolated dextrocardia
- b) Dextrocardia with situs inversus

Course of the Circulation

No alteration in hemodynamics unless an associated malformation is present

Clinical Features

Asymptomatic

Apex impulse visible on the right

Heart tones heard best over right anterior chest

Liver edge may be palpable in left upper quadrant

Electrocardiogram

Lead I mirror image of normal

Lead II resembles normal lead III

Lead III resembles normal lead II

Radiological Studies

Mirror image of normal

Fluorocopy of value in establishing position of liver and stomach

Prognosis

a) Isolated dextrocardia—guarded

b) Dextrocardia with situs inversus—good

Treatment

No treatment required unless an associated malformation is present

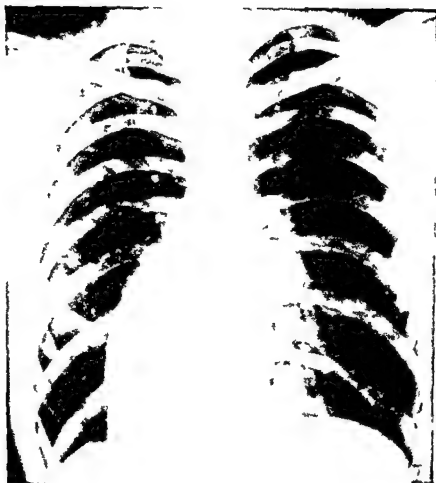


Fig 1

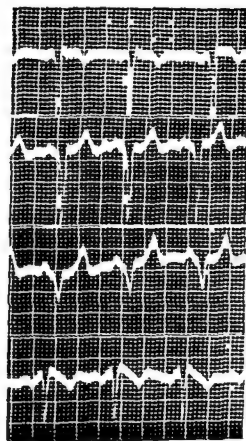


Fig 2

Isolated Dextrocardia (Adult)

Fig 1 A P roentgenogram of the chest showing typical dextrocardia

Fig 2 E K C Notice that lead I is a mirror image of the normal tracing. The Q R S complex, P waves and T waves are inverted in lead I. The P waves and T waves are upright in lead III.

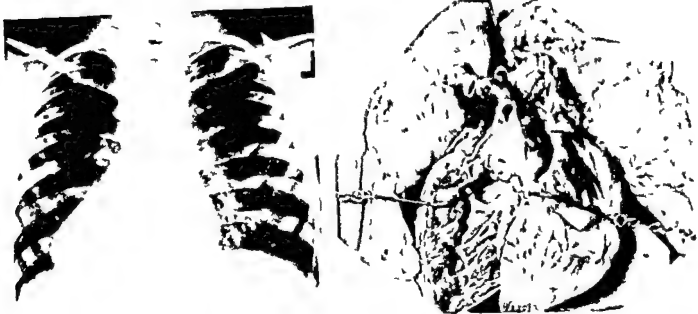


Fig 3

Tetralogy of Fallot and Situs Inversus
(1 year 9 years)

Fig 3 A P roentgenogram of the chest showing dextrocardia associated with tetralogy of Fallot

Fig 4 Heart and lungs Notice the aortic arch curving to the right the left-sided position of the anatomical right ventricle and the large ventricular septal defect

Fig 5 Heart and lungs from the same case Notice the small pulmonary artery to the right of the aorta

Fig 6 EKG Notice that the changes represent a composite of those produced by dextrocardia and the ones due to tetralogy of Fallot The QP S complexes are upright in lead I and inverted in lead III The I waves and T waves are inverted in lead I and upright in leads II and III The P waves are peaked in lead II



Fig 5

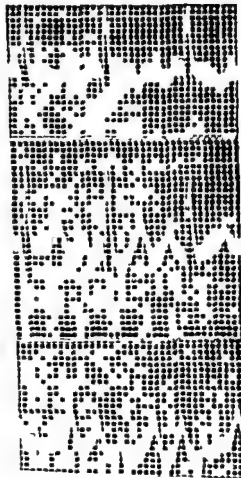


Fig 6

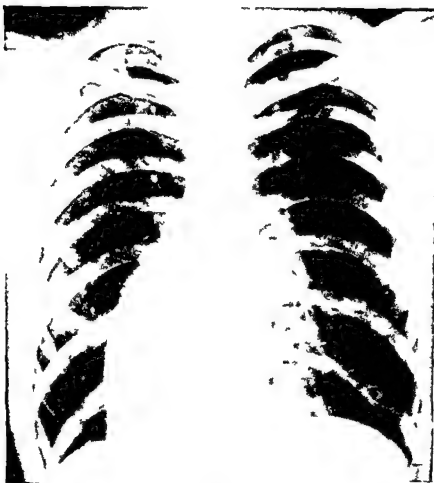


Fig 1

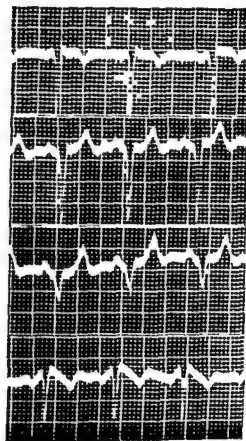


Fig 2

Isolated Dextrocardia

(Adult)

FIG. 1 AP roentgenogram of the chest showing, typical dextrocardia

FIG. 2 ECG Notice that lead I is a mirror image of the normal tracing. The QRS complexes, P waves and T waves are inverted in lead I. The P waves and T waves are upright in lead III.

2

Defects of the Cardiac Septa

Pathology

Defects of the auricular septum include

- a) Persistent foramen ovale
- b) Defects at the site of the foramen ovale
- c) Defects in the region of the ostium primum
- d) Complete absence of the auricular septum

Defects of the ventricular septum

- a) Defects in the upper membranous septum
- b) Defects in the muscular septum
- c) Complete absence of the ventricular septum

Interventricular Syndrome consists of an auricular septal defect associated with a mitral valve stenosis (Plate I)

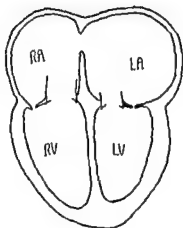


PLATE I

Interventricular syndrome (auricular septal defect associated with mitral valve stenosis)

Course of the Circulation

- a) Patent foramen ovale—right to left shunt only
- b) Other auricular defects—left to right shunt—reversing at times of stress
- c) Ventricular septal defects—left to right shunt—reversing if pulmonary hypertension develops
- d) Interventricular Syndrome—usually a large left to right shunt resulting in right sided cardiac enlargement and enlargement of pulmonary artery

Defects of the Auricular Septum

Clinical Features

- Poor physical growth and development
- Transitory cyanosis in infancy
- Repeated respiratory infections
- Cardiac enlargement
- Narrow pulse pressure
- Systolic murmur

2

Defects of the Cardiac Septa

Pathology

Defects of the auricular septum include

- Persistent foramen ovale
- Defects at the site of the foramen ovale
- Defect in the region of the ostium primum
- Complete absence of the auricular septum

Defects of the ventricular septum

- Defects in the upper membranous septum
- Defect in the muscular septum
- Complete absence of the ventricular septum

Lutembacher's Syndrome consists of an auricular septal defect associated with a mitral valve stenosis (Plate 1)

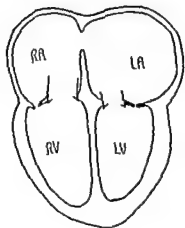


PLATE 1

Lutembacher's Syndrome (auricular septal defect associated with mitral valve stenosis)

Course of the Circulation

- Patent foramen ovale—right to left shunt only
- Other auricular defects—left to right shunt—reversing at times of stress
- Ventricular septal defects—left to right shunt—reversing if pulmonary hypertension develops
- Lutembacher's Syndrome—usually a large left to right shunt resulting in right sided cardiac enlargement and enlargement of pulmonary artery

Defects of the Auricular Septum

Clinical Features

- Poor physical growth and development
- Transitory cyanosis in infancy
- Repeated respiratory infections
- Cardiac enlargement
- Narrow pulse pressure
- Systolic murmur

Electrocardiogram

- Right axis deviation
- Right ventricular preponderance
- Peaked P waves
- Prolonged P R interval

Radiological Studies

- Right auricular and right ventricular enlargement
- Prominent pulmonary artery segment
- Increased vascular lung markings
- Hilar pulsations noted on fluoroscopy

Laboratory

- Microcytic anemia
- Increased right auricular pressure
- Increased O₂ saturation in right auricle

Prognosis

- Poor, may reach adult life but physical activity is usually restricted

Treatment

- Prevention of respiratory infection
- Treatment of cardiac failure
- Surgical closure of defects possible but not completely evaluated



Fig 7

Auricular Septal Defect

Fig 7 Large auricular septal defect in an adult female

Fig 8 A P roentgenogram of the chest in a two-and-a-half year old child with a large auricular septal defect. There is cardiac enlargement and heavy vascular markings in both lung fields.



Fig 8

Lutembacher's Syndrome

Except for the huge size of the pulmonary artery on the roentgenograms the clinical features of a Lutembacher's Syndrome are quite similar to those of an auricular septal defect



Fig 9



Fig 10

Lutembacher's Syndrome

(Age 12 years)

Fig 9 Heart from a case of Lutembacher's syndrome showing a large auricular septal defect (ostium primum) and a thickened and deformed mitral valve

Fig 10 AP roentgenogram of the chest. The picture reveals a large pulmonary conus and heavy vascular lung markings

Fig 11 EKG showing right axis deviation and delayed AV conduction. The QRS complexes are inverted in lead I and upright in lead II. The P-R interval is 0.20 sec

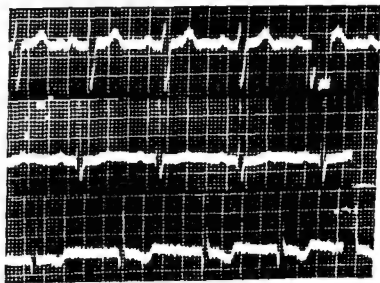
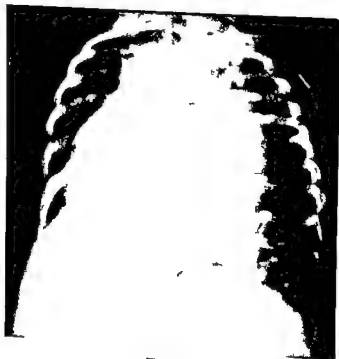


Fig 11

Lutembacher's Syndrome*(Age 7 years)*

Fig 12 A P roentgenogram of the chest from another case of Lutembacher's syndrome. The picture is similar to Fig 10 but shows a larger right atrium.

Fig 13 L A O roentgenogram of the chest from the same case. Notice the very prominent right ventricle.

**Fig 12****Fig 13**

Defects of the Ventricular Septum

Clinical Features

- Usually asymptomatic
- Growth and development normal
- Heart normal size
- Systolic murmur—3rd and 4th left ICS close to sternum
- Systolic thrill—3rd and 4th left ICS close to sternum

Electrocardiogram

- Usually normal

Radiological Studies

- Heart normal in size and configuration

Laboratory

- O₂ saturation of blood in the right ventricular outflow tract exceeds that of the right ventricle or right auricle

Prognosis

- Excellent, unless pulmonary hypertension develops

Treatment

- Surgical closure possible but not completely evaluated

Ventricular Septal Defect

Fig 14 Defect in the membranous portion of the ventricular septum. The anomaly was found incidentally at autopsy in an adult male

Fig 15 Defect in the muscular portion of the ventricular septum in an infant

Ventricular Septal Defect with Deformity of Aortic Valve

(8 year old male)

Fig 16 Opened left ventricle showing the ventricular septal defect, the deformity of the right aortic leaflet which overlies the septal defect and bacterial endocarditis involving the upper septum and the aortic valve

Fig 17 Close up of the aortic valve and the ventricular septal defect. Patient had a continuous murmur at the base of the heart and high pulse pressure

Fig 18 Opened right ventricle showing hypertrophy and ventricular septal defect

Fig 19 A P radiograph of the chest showing cardiac hypertrophy, prominence of the pulmonary artery and increased pulmonary vascular markings



Fig 14



Fig 15



Fig 16



Fig 17



Fig 18



Fig 19

Common Atrioventricular Ostium

Pathology

Incomplete septation of the atrioventricular canal resulting in

- a) Defect in the lower portion of the auricular septum
- b) Defect in the upper portion of the ventricular septum
- c) Common atrioventricular valve

Course of the Circulation

Complete pooling of blood

Clinical Features

Frequently associated with mongolism

Cyanosis slight

Cardiac enlargement

Systolic murmur

Systolic thrill

Electrocardiography

No diagnostic pattern

Usually right axis deviation

Radiological Studies

Cardiac enlargement

Increased vascular lung markings

Laboratory

Decreased arterial O₂ saturation

Prognosis

Average length of life ten to twelve years

Treatment

Prevention of respiratory infections

Management of cardiac failure

Common Atrioventricular Ostium

(6 month old male)

Fig 20 Common atrioventricular ostium. Notice the opened right ventricle, the large defect in the lower part of the auricular septum, and the common valve with leaflets crossing the midline through the septal defect.



Fig 20

Fig 21 A P roentgenogram of the chest from the same case as the specimen in Fig 20. The heart is enlarged to right and left and the pulmonary vascular markings are heavy.



Common Atrioventricular Ostium

Pathology

Incomplete septation of the atrioventricular canal resulting in

- a) Defect in the lower portion of the auricular septum
- b) Defect in the upper portion of the ventricular septum
- c) Common atrioventricular valve

Course of the Circulation

Complete pooling of blood

Clinical Features

Frequently associated with mongolism

Cyanosis slight

Cardiac enlargement

Systolic murmur

Systolic thrill

Electrocardiography

No diagnostic pattern

Usually right axis deviation

Radiological Studies

Cardiac enlargement

Increased vascular lung markings

Laboratory

Decreased arterial O₂ saturation

Prognosis

Average length of life ten to twelve years

Treatment

Prevention of respiratory infections

Management of cardiac failure



Fig 22



Fig 23



Fig 24

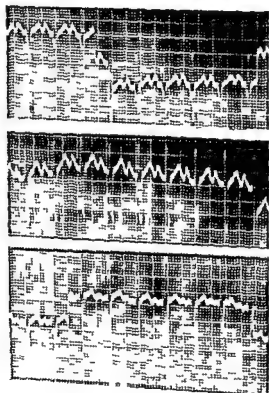


Fig 25

Common Atrioventricular Ostium

(16 month old male)

Fig 22 Heart from a boy with common atrioventricular ostium. Picture shows the left atrium and left ventricle with a large septal defect. Notice the common A V valve with some leaflets running through the septal defect.

Fig 23 Common atrioventricular orifice with six valvular leaflets: two common leaflets (across midline), one leaflet in the left ventricle, and three in the right ventricle (two of these are rudimentary).

Fig 24 A P roentgenogram of chest from the same case shows marked cardiac enlargement and congestion in the lung fields.

Fig 25 E K G showing tachycardia, prominence of P_1 and P_2 and a diphasic P_1 , also right axis deviation.

Cor Triloculare

Pathology (Plates 3-4)

Complete absence of auricular septum—*cor triloculare biven- triculare*

Complete absence of ventricular septum—*cor triloculare biatriatum*

There are usually other associated cardiovascular anomalies

Course of the Circulation

Complete pooling within the common chamber

Clinical Features

Cyanosis from early infancy

Cardiac enlargement

Poor growth and development

Systolic murmur

Electrocardiogram

Right axis deviation

Abnormal P waves

Delayed A-V conduction

Radiological Studies

Cardiac enlargement

Lung fields appear congested

Laboratory

Arterial O₂ saturation decreased

Prognosis

Poor—usually expire in early infancy

Treatment

None

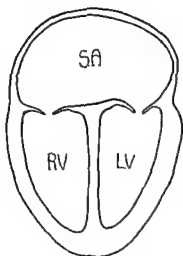


PLATE 3

Cor triloculare biven- triculare (a three chamber heart with a single auricle and two ventricles)

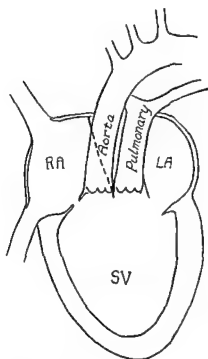


PLATE 4

Cor triloculare biatriatum (a three chamber heart with two auricles and a single ventricle)

Cor Biloculare

Pathology (Plate 2)

Complete absence of auricular and ventricular septa

Common atrioventricular ostium

Occasionally associated lesions are transposition of the great vessels and tricusus uteriosus

Course of the Circulation

Complete pooling within the common auricle and ventricle



Fig 26

Cor Biloculare

Fig. 26 Heart specimen from an infant with cor biloculare. Notice the complete absence of auricular and ventricular septa and the common atrioventricular valve.

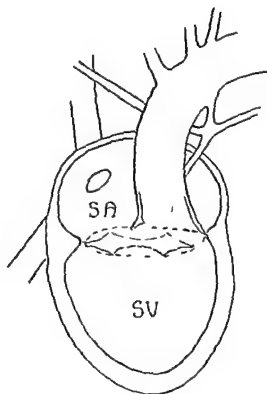


PLATE 2

Cor biloculare (a two chamber heart a single auricle and single ventricle)

Cor Triloculare

Pathology (Plates 3-4)

Complete absence of auricular septum—*cor triloculare biven-
triculare*

Complete absence of ventricular septum—*cor triloculare
biatriatum*

There are usually other associated cardiovascular anomalies

Course of the Circulation

Complete pooling within the common chamber

Clinical Features

Cyano is from early infancy

Cardiac enlargement

Poor growth and development

Systolic murmur

Electrocardiogram

Right axis deviation

Abnormal P waves

Delayed A-V conduction

Radiological Studies

Cardiac enlargement

Lung fields appear congested

Laboratory

Arterial O₂ saturation decreased

Prognosis

Poor usually expire in early infancy

Treatment

None

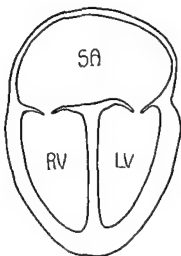


PLATE 3

Cor triloculare biventriculare (a three chamber heart—a single auricle and two ventricles)

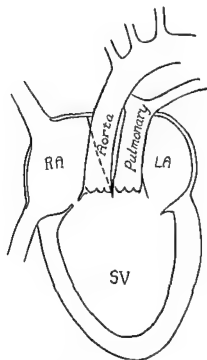


PLATE 4

Cor triloculare biatriatum (a three chamber heart with two auricles and a single ventricle)



Fig. 27

Cor Triloculare Biventriculare

(Age 4 months)

Fig. 27 Heart from a case of cor triloculare biventriculare. Notice the complete absence of atricular septum, and the small ventricular septal defect. Notice also the common atrioventricular valve.

Fig. 28 A P roentgenogram from same case of cor triloculare biventriculare as shown in Fig. 27. There is a large globular heart and very vascular lung fields.



Fig. 28

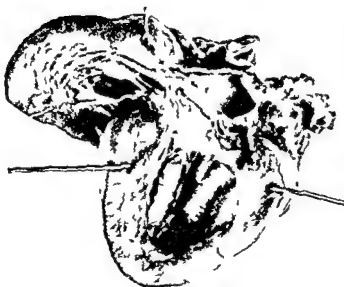


Fig 29

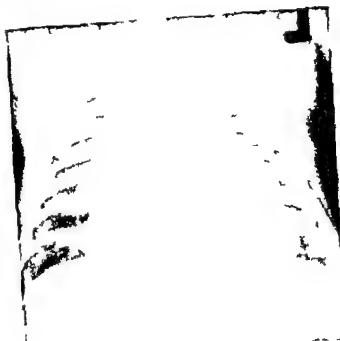


Fig 30

Cor Triloculare Batriatum

(1gc 8 days)

Fig 29 Heart from a case of cor triloculare batriatum Notice the single ventricle with the four cardiac valves opening into it
 Fig 30 A roentgenogram from same case of cor triloculare batriatum as shown in Fig 29 It shows cardiac enlargement and increased pulmonary vascular markings
 Fig 31 Section of lung from same case of cor triloculare batriatum Notice the thick walled blood vessels especially in the subpleural region

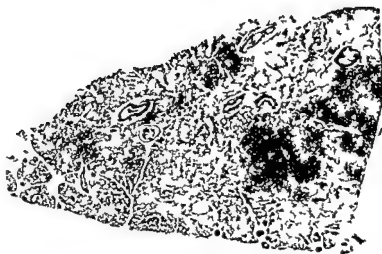


Fig 31



Fig 32

Cor Triatriatum*(6 months old female)*

Fig 32 Superior view of the heart showing the accessory left atrium. The probes mark the points of entry of all four pulmonary veins. A short probe is directed down toward the main left atrium.

Fig 33 Left ventricle, mitral valve and left atrium. The only communication between the left atrium and the accessory atrium is through a narrow opening as shown by probe.



Fig 33

3

Valvular Anomalies

Tricuspid Valve Atresia

Pathology (Plate 5)

- Complete obliteration of tricuspid orifice
- Hypoplasia of right ventricle
- Hypoplasia of pulmonary valve and artery
- Associated defects of the auricular and ventricular septa
- Patent ductus arteriosus

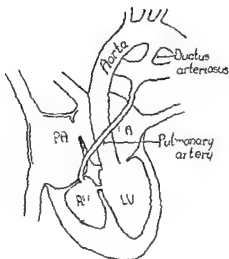


PLATE 5

Tricuspid atresia with under developed right ventricle

Course of the Circulation

- Right atrium → left atrium → left ventricle → aorta
- systemic circulation
- patent ductus or bronchial arteries to the lung

Clinical Features

- Cyano is from birth a common finding
- Heart normal size
- Systolic murmur
- Enlarged liver
- Pulsations at liver margin present on occasion

Electrocardiogram

- Usually left axis deviation
- Left ventricular preponderance
- Peaked P waves
- (Fig 3 is an exception)

Radiological Findings

- Decreased bronchovascular markings
- Heart size normal
- Concavity in the region of the pulmonary conus P A view
- Right cardiac border has double convexity P A view
- Shadow cast by right ventricle absent in I A O view

Laboratory

Arterial O₂ unsaturation
Hemoconcentration

Prognosis

Poor
Average length of life 4-6 months

Treatment

Surgical creation of an auricular defect or the creation of an extracardiac shunt is of benefit



Fig 34

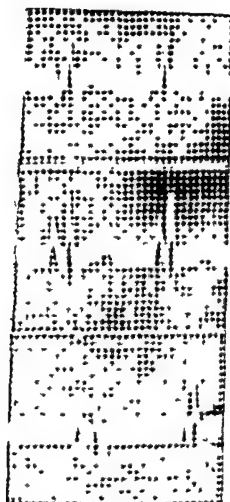


Fig 35

**Tricuspid Atresia and Pulmonary
Valve Atresia**
(Age 4 months)

Fig 34 Heart in a case of tricuspid atresia. Notice the dilated right atrium and the atrioventricular orifice.

Fig 35 ECG showing peaked P wave in leads II and III but no evidence of left axis deviation.

Fig 36 Pulmonary artery and valve in the same case as Fig 34. Notice the atrioventricular pulmonary valve orifice.

Fig 37 A P roentgenogram of the heart in the same case. The heart is of normal size and shows slight convexity in the region of the pulmonary conus and a double convexity of the right cardiac border. The lung fields are avascular.

Fig 38 I A O roentgenogram of chest. Notice the normal size of the left (posterior) ventricle and the small size of the right (anterior) ventricle.



Fig 36



Fig 37



Fig 38



Fig 39



Fig 10

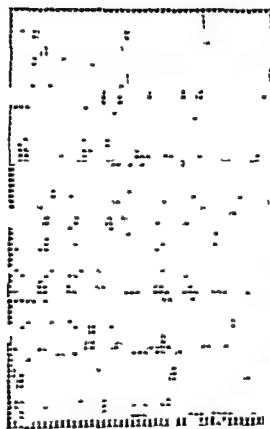


Fig 11

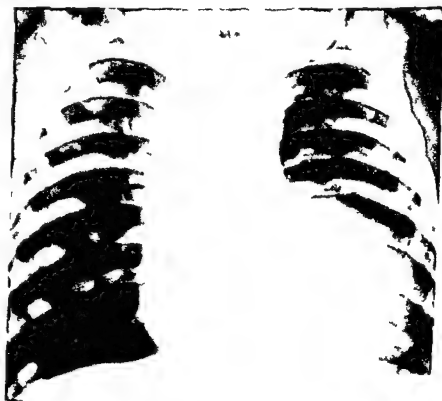


Fig 12

Tricuspid Atresia
(with absence of auricular septum)

(Age $2\frac{1}{2}$ years)

Fig 39 Heart from male with tricuspid atresia Notice the absence of auricular septum and the atretic tricuspid orifice

Fig 40 Anterior view of the same heart as in Fig 39 Notice the small pulmonary artery and the calcified nodular thrombus in the infundibulum

Fig 41 E K G Notice the peaked P waves in leads I and II with evidence of left axis deviation The Q R S complexes are upright in lead I and inverted in lead III

Fig 42 A P roentgenogram of the chest from a $2\frac{1}{2}$ year old male with tricuspid atresia and complete absence of auricular septum Notice the left sided enlargement of the heart the concavity in the region of the pulmonary conus and the double convexity in the right border The lung fields are avascular

Fig 43 Sections of aorta and pulmonary artery The longer section is from the pulmonary artery At $2\frac{1}{2}$ years the two sections should be of about equal thickness Elastic tissue stain ($\times 15$)

Fig 44 Section of lung Notice the thin walled pulmonary arteries and their relatively small size Elastic tissue stain ($\times 15$)

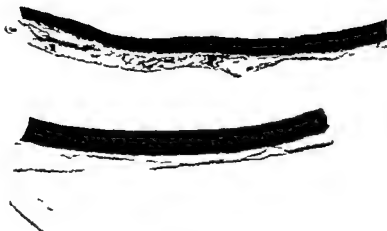


Fig 43

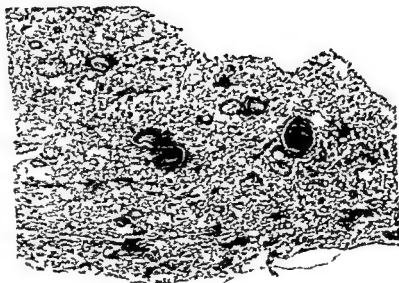


Fig 44

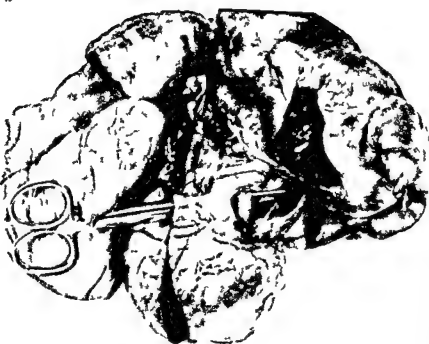


Fig 45



Fig 46

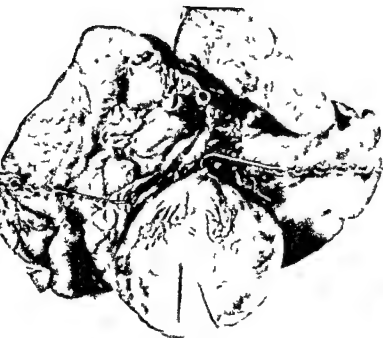


Fig 47



Fig 48

Tricuspid Atresia
(with auricular septal defect)

(Age 10 years)

Fig. 45 Heart and lungs from a 10 year old colored male with tricuspid atresia. The scissors pass through a large defect in the auricular septum. Notice the atresia of the tricuspid orifice.

Fig. 46 Anterior view of the same heart. Notice the prominent left ventricle, the small pulmonary artery, and the undeveloped right ventricle which consists only of a small outflow (infundibular) portion. The probe passes through a small ventricular septal defect.

Fig. 47 View of the heart showing small hypoplastic pulmonary artery.

Fig. 48 Descending thoracic aorta showing the intercostal arteries and some large bronchial arteries.

Fig. 49 Section of the aorta and pulmonary artery. Notice the thin walled pulmonary artery which is the longer of the two specimens shown.

Fig. 50 Section of the lung showing small and thin walled pulmonary arteries. Elastic tissue stain ($\times 15$).



Fig. 49

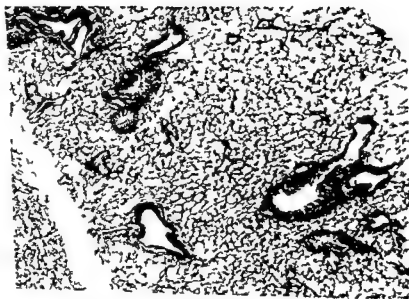


Fig. 50

Pulmonary Valve Atresia

Pathology (Plate 6)

Pulmonary valve obliterated
 Pulmonary artery reduced in size
 A patent ductus arteriosus frequently present
 Auricular septal defect
 Large dilated right auricle
 Small and thick walled right ventricle

Course of the Circulation

Right auricle → left auricle → left ventricle
 aorta → systemic circulation
 ↙
 patent ductus or enlarged bronchial arteries to
 the lungs

Clinical Features

Cyanosis and dyspnea
 Progressive cardiac enlargement
 Liver enlarged

Electrocardiography

Right axis deviation
 Peaked P waves are common

Radiological Studies

Marked cardiac enlargement (right auricle)
 Avascularity of lung fields

Laboratory

Arterial O₂ unsaturation
 Hemoconcentration
 Increased right ventricular and right auricular pressure revealed by catheterization

Prognosis

Poor
 Death at 4-6 months

Treatment

Pulmonary valvulotomy of value if the pulmonary artery is not markedly hypoplastic

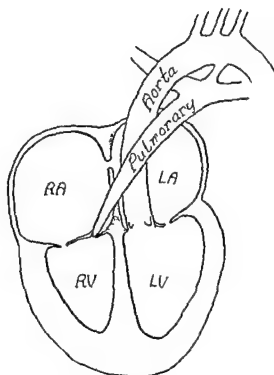


PLATE 6
 Pulmonary valve atresia

Pulmonary Valve Stenosis

Pathology

Fusion of valvular cusps into a small "volcano like" structure with the narrow valvular orifice as the crater
 Post stenotic dilation of the pulmonary artery
 Right ventricular hypertrophy
 Often an associated atricular septal defect or patent foramen ovale
 Intact ventricular septum

Course of the Circulation

In most instances the hypertrophied right ventricle forces blood through the stenotic orifice
 If an atricular septal defect is present the flow is from right to left

Clinical Features

Variable depending upon absence or presence of atricular defect
 Dyspnea an outstanding symptom
 Cyanosis if atricular defect is present
 Heart normal in size
 Systolic murmur
 Systolic thrill
 Decreased or absent P

Electrocardiogram

Right axis deviation
 Right ventricular preponderance
 High peaked P waves

Radiological Studies

Heart normal size or slightly enlarged
 Vascular lung markings normal or decreased
 Prominent pulmonary conus (post stenotic dilation)
 Angiocardiography of value in demonstrating presence of an atricular septal defect

Laboratory

There is usually arterial O₂ unsaturation
 Catheterization reveals an increased right ventricular pressure and a normal or low pulmonary artery pressure

Prognosis

In general good

Treatment

Pulmonary valvulotomy

Pulmonary Valve Atresia

Pathology (Plate 6)

- Pulmonary valve obliterated
- Pulmonary artery reduced in size
- A patent ductus arteriosus frequently present
- Auricular septal defect
- Large dilated right auricle
- Small and thick walled right ventricle

Course of the Circulation

Right auricle → left auricle → left ventricle
 aorta → systemic circulation

↙ patent ductus or enlarged bronchial arteries to
 the lungs

Clinical Features

- Cyanosis and dyspnea
- Progressive cardiac enlargement
- Liver enlarged

Electrocardiography

- Right axis deviation
- Peaked P waves are common

Radiological Studies

- Marked cardiac enlargement (right auricle)
- Avascularity of lung fields

Laboratory

- Arterial O₂ unsaturation
- Hemoconcentration
- Increased right ventricular and right auricular pressure revealed by catheterization

Prognosis

- Poor
- Death at 4-6 months

Treatment

- Pulmonary valvulotomy of value if the pulmonary artery is not markedly hypoplastic

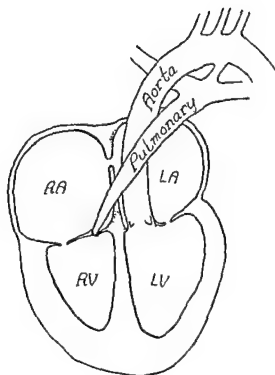


PLATE 6
 Pulmonary valve atresia



Fig 53

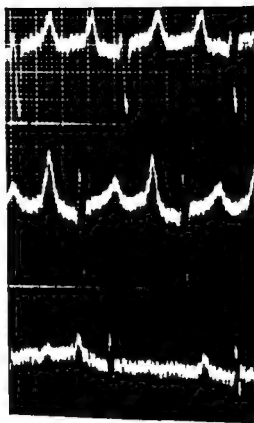


Fig 54



Fig 51

Pulmonary Valve Atresia

(Age 8 weeks)

Fig 51 Pulmonary valve atresia Notice the membranous formation which obliterates the pulmonary valvular orifice. The right ventricle is small but very thick walled. The right atrium is enormously dilated. The ductus arteriosus is patent.

Fig 52 Posterior view of the heart. Notice the dilated right atrium, the small left atrium, and the auricular septal defect.

Fig 53 A P roentgenogram of the chest. Notice the marked cardiac enlargement to the right and left. The large right atrium can be seen.

Fig 54 E K G showing peaked P waves in leads I and II. There is right axis deviation. The Q R S complexes are inverted in lead I and upright in lead III.



Fig 52

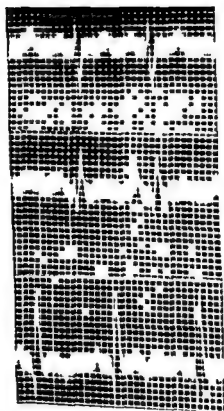


Fig 57



Fig 58



Fig 55

Pulmonary Valve Stenosis
(with auricular septal defect)

(25 year old female)

Fig 55 Pulmonary valve stenosis. Pulmonary valve seen from above. Notice the small circular opening and the position of the commissures. Notice also the marked dilatation of the pulmonary artery.

Fig 56 A P roentgenogram of the chest. Notice the large pulmonary artery and the vascular lung fields.

Fig 57 E K G showing a marked right axis deviation and peaked P waves in lead II. The QRS complexes are inverted in lead I and upright in lead III.

Fig 58 Sections of aorta and pulmonary artery. Notice the extremely thin wall of the pulmonary artery. Elastic tissue stain ($\times 15$).





Fig 62

Pulmonary Infundibular Stenosis*(69 year old female)*

Fig 62 Notice the narrow opening in the outflow tract of the right ventricle. The ventricular septum is intact. The pulmonary valve is unchanged.



Fig 59

Pulmonary Valve Stenosis
(with an intact auricular septum)

(Age 8 months)

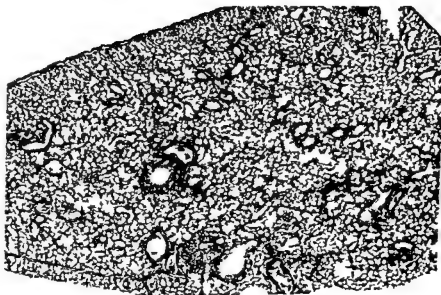
Fig 59 Heart and lungs from an 8 month old child with pulmonary valve stenosis. Notice the typical deformity of the pulmonary valve with a round crater in the center. There is moderate pulmonary dilatation, and marked hypertrophy of the right ventricle and right atrium.

Fig 60 Section of aorta and pulmonary artery. Notice the marked thinness of the pulmonary artery wall. Elastic tissue stain ($\times 15$).

Fig 61 Section of lung revealing small and thin walled pulmonary arteries. Elastic tissue stain ($\times 15$).



Fig 60



Aortic Valve Atresia

Pathology (Plate 8)

Aortic valve atresia
 Hypoplasia of left ventricle
 Hypoplastic aorta
 Right auricular-right ventricular enlargement
 Pulmonary artery enlarged
 Patent ductus arteriosus

Course of the Circulation

Left ventricular and left auricular blood → right auricle
 → right ventricle → pulmonary artery → patent ductus
 arteriosus → aorta

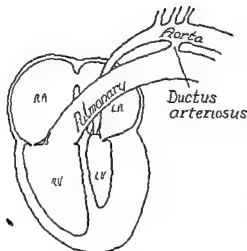


PLATE 8
 Aortic valve atresia

Clinical Features

Protruding cyanosis and cardiac enlargement
 Systolic murmur

Electrocardiogram

Right axis deviation
 Peaked P waves

Radiological Studies

Cardiac enlargement
 Normal or increased vascular lung markings

Laboratory

Arterial O_2 saturation is decreased
 Hemoconcentration
 O_2 content in right auricular blood sample increased

Prognosis

Poor
 Most infants with this malformation die within the first few weeks of life

Treatment

None

4

Anomalies of Septation of Truncus Arteriosus

Persistent Truncus Arteriosus

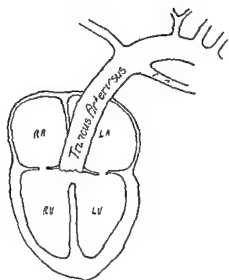


PLATE 9

Complete truncus arteriosus

Pathology

There is only one vessel arising from the base of the heart. This single vessel is the origin of the coronary, pulmonary and systemic vessels.

A defect in the membranous portion of the ventricular septum.

There are frequently four cusps in the valve of the vessel arising from the base of the heart.

Plates 9 and 10 illustrate complete and partial truncus arteriosus.

Course of the Circulation

The ventricular septal defect and the common trunk for the pulmonary and systemic circulations result in marked pooling of arterial and venous blood.

Circulation to the lungs is by way of pulmonary vessels arising from the truncus or by the way of bronchial vessels.

Clinical Features

Cyanosis moderate

Heart normal size or slightly enlarged

Systolic murmur

Clear cut second sound

Electrocardiogram

Right axis deviation



Fig. 63

Aortic Valve Atresia

(6 months old male)

Fig. 63 Large and thick walled pulmonary artery, hypoplastic aorta and patent ductus arteriosus. Notice the large right atrium and right ventricle.



Fig. 64

Fig. 64 Posterior view of the heart showing marked hypoplasia of the left atrium and left ventricle, a patent foramen ovale and hypertrophy and dilatation of both right-sided chambers.

Persistent Truncus Arteriosus Partial Type

(Age 8 days)

Fig. 65 Partial truncus arteriosus communis persistens showing incomplete aortic pulmonary septum left coronary ostium and short pulmonary trunk with its two branches

Fig. 66 AP roentgenogram of the chest. Notice marked enlargement of cardiac outline to left absence of pulmonary conus and small size of aortic knob

Fig. 67 LAO roentgenogram of the chest. Notice the large left ventricle. The anterior border of the right ventricle is only slightly more prominent than the border of the common trunk



Fig. 65



Fig. 66

Radiological Studies

Right and left ventricular enlargement

When cyanosis is minimal, marked hilar pulsations are usually present

When cyanosis is marked the lung fields are avascular

Laboratory

O₂ saturation of the arterial blood decreased

Hemoconcentration may be present

Prognosis

Variable—length of life depends on blood supply to the lungs

Treatment

None

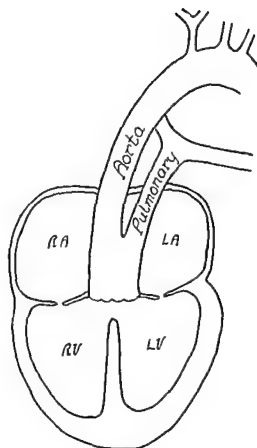


PLATE 10

Partial transposition arteriosus



Fig 71

**Persistent Truncus Arteriosus
Complete Type**

(Age 10 months)

Fig 71 Complete truncus arteriosus. Notice displacement of arterial trunk to the right fourth ribs cupping single large coronary ostium and large pulmonary arteries coming directly off the truncus.

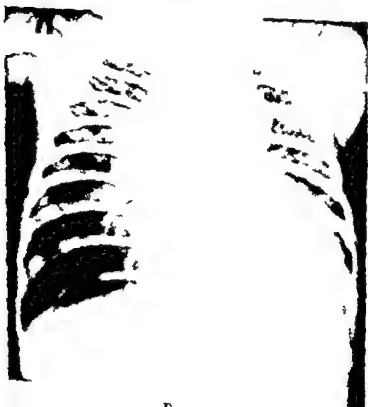


Fig 72 AP roentgenogram of the chest. Notice marked enlargement of cardiac shadow to left absence of pulmonary conus and enormous aortic knob.



FIG. 68

Persistent Truncus Arteriosus, Partial Type

(Age 6 weeks)

Fig. 68 Partial truncus arteriosus communis persists. Notice incomplete aortic pulmonary septum between pulmonary trunk and ascending aorta. The ventricular septal defect is plainly seen.

Fig. 69 AP roentgenogram of the chest. Notice the marked enlargement of the cardiac outline to the left and the absence of the pulmonary conus, also the small size of the aortic knob.

Fig. 70 LAO roentgenogram of the chest. Notice the large left ventricle. The anterior border of the right ventricle is only slightly more prominent than the border of the common trunk.



FIG. 70

Solitary Aortic and Pulmonic Trunk

Pathology (Plates 11, 12)

Eccentric septation of the truncus arteriosus
 Solitary aortic trunk with pulmonary atresia—or
 Solitary pulmonic trunk with aortic atresia
 Ventricular septal defect

Course of the Circulation

Solitary aortic trunk—blood from both ventricles enters the solitary arterial trunk and the blood reaches the lungs through a patent ductus

Solitary pulmonic trunk—blood from both ventricles enters the pulmonic trunk, pulmonary vessels arise from the pulmonic trunk and the ductus arteriosus continues as the descending aorta

Clinical Features

Similar to that of a truncus arteriosus or partial truncus arteriosus

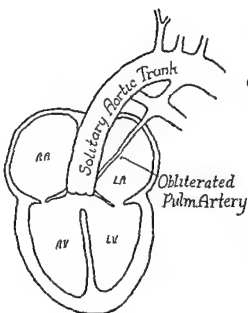


PLATE 11
 Solitary aortic trunk

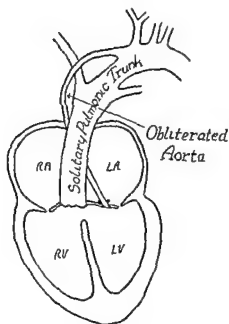


PLATE 12
 Solitary pulmonic trunk

Persistent Truncus Arteriosus, Partial Type

(Age 7 days)

Fig. 73 "Partial" truncus arteriosus communis. Notice the four "fleshy" cusps in the valve, the incomplete aortic pulmonary septum, and the ventricular septal defect.



Fig. 73

Persistent Truncus Arteriosus with Right Aortic Arch and Right Descending Aorta

(Age 3 weeks)

Fig. 74 "Partial" truncus arteriosus communis. Notice the incomplete aortic pulmonary septum and the right sided aortic arch.



Aortic Septal Defect

Pathology (Plate 13)

Incomplete septation of the truncus arteriosus

Both great vessels arise from the heart in a normal fashion

Communication between pulmonary artery and aorta close to the base of the heart

Course of the Circulation

Flow through aortic septal defect is left to right, that is from aorta to pulmonary artery

Clinical Features

Growth and development may be retarded or normal

Peripheral signs of aortic insufficiency

Continuous roaring murmur over the base of the heart

Thrill over base of heart

Electrocardiogram

Usually normal

On occasion left ventricular preponderance

Radiological Studies

Vascular lung markings increased

Hilar pulsations observed on fluoroscopy

Evidence of left ventricular enlargement

Laboratory

Arterial O_2 saturation normal

Catheterization reveals O_2 content of blood from pulmonary artery exceeds that of the right ventricle

Pulmonary pressure may be increased

Prognosis

Good

Treatment

Surgical closure of the defect

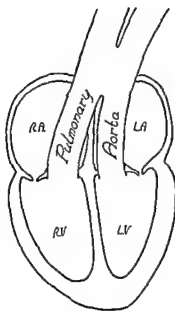


PLATE 13
Aortic septal defect



Fig 75

Solitary Aortic Trunk with Pulmonic Atresia*(Age 8 days)*

Fig 75 Heart from a case of solitary aortic trunk (pseudotruncus?) Notice the large aorta and the atretic pulmonary artery. Notice also the patent ductus arteriosus which communicates with the arterial branches to both lungs

5

Transposition Complexes

Eisenmenger's Complex

Pathology (Plate 14)

- Right ventricular hypertrophy
- Ventricular septal defect
- Dextro-position of the aorta
- Pulmonary artery large and dilated

Course of the Circulation

The dextro-positioned aorta receives blood from both ventricles. The enlarged pulmonary artery results in an increased flow and increased pressure in the pulmonary circulation.

Clinical Features

- Onset of cyanosis in late childhood
- Slight cardiac enlargement
- Basal systolic murmur and thrill
- Hemoptysis is frequent

Electrocardiogram

- Right axis deviation
- Right ventricular preponderance
- Peaked P waves

Radiological Studies

- Contour of the heart varies with the size of the pulmonary artery
- If pulmonary artery is enlarged, contour resembles that seen in cases of Lutembacher's syndrome
- If pulmonary artery is not enlarged, contour resembles that of a Tetralogy of Fallot

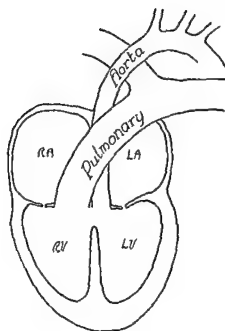


PLATE 14
Eisenmenger's complex

Radiological Studies

Heart normal size

Concave pulmonary contour

Upturned apex

25% right aortic arch

Angiascular lung fields

Angiocardiography of value in determining position of aorta

Laboratory

Arterial O₂ saturation markedly reduced

Hemoconcentration

Cardiac catheterization reveals increased right ventricular pressure catheter may pass through ventricular defect into aorta

Prognosis

Compatible with long life however physical activity may be moderately or markedly restricted

Treatment

Surgical designed to increase blood flow to lungs

Blalock-Taussig subclavian artery pulmonary artery anastomosis

Potts-Smith direct aorta pulmonary artery anastomosis

Brock Procedure direct attack on the pulmonary stenosis

The lung fields are quite vascular, hilar pulsations conspicuous

Angiocardiography results in simultaneous filling of the right ventricle, enlarged pulmonary artery and aorta

Laboratory

Arterial O₂ unsaturation

Catheterization reveals an increased right ventricular and pulmonary artery pressure

Prognosis

Once cyanosis develops the prognosis is guarded

Treatment

No surgical treatment

Tetralogy of Fallot

(Illustrations begin page 56)

Pathology (Plate 15)

Pulmonary stenosis valvular or infundibular

Dextroposition of the aorta

Right ventricular hypertrophy

Ventricular septal defect

Course of the Circulation

The dextroposed aorta receives blood from both ventricles
Decreased pulmonary flow as a result of the pulmonary stenosis

Bronchial arteries become important in blood supply to lungs

Clinical Features

Cyanosis and clubbing

Heart normal size

Left anterior chest deformity

Basal systolic murmur and thrill

Electrocardiogram

Right axis deviation

Right ventricular preponderance

Peaked P waves

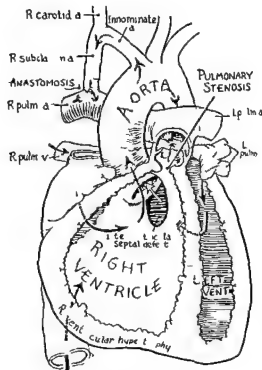


PLATE 15

Tetralogy of Fallot with a Blalock-Taussig shunt

Eisenmenger Complex
(with right sided aortic arch and descending aorta)

(Age 8 months)



Fig. 80

Fig. 76 Eisenmenger complex. Notice the large size of the pulmonary artery. The arch of the aorta and the descending portion are to the right of the trachea.

Fig. 77 Eisenmenger complex. Same case showing the opened left ventricle and the high ventricular septal defect.

Fig. 78 AP roentgenogram of the chest. Notice the cardiac enlargement and the heavy vascular markings of the lungs.

Fig. 79 EKG showing right axis deviation, peaked P waves in lead II and diphasic P waves in lead III. The QRS complex is inverted in lead I and upright in lead III.

Fig. 80 Eisenmenger complex. Section of aorta and pulmonary artery (longest section). Notice the hypertrophy of the pulmonary artery which is thicker than the aorta. Elastic tissue stain ($\times 15$).

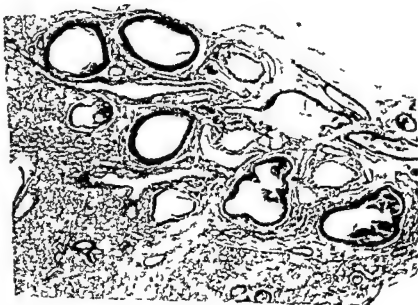


Fig. 81 Eisenmenger complex. Section of lung showing the large and thick walled pulmonary arteries. Elastic tissue stain ($\times 10$).

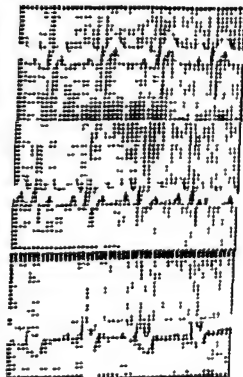
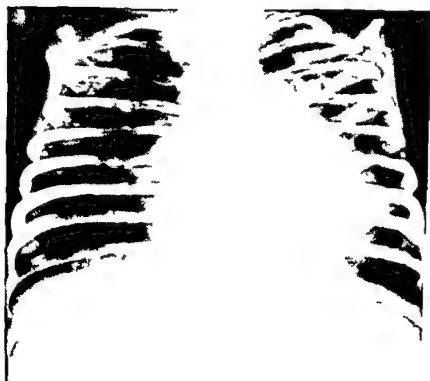
Fig. 81



Fig 76



Fig 77



Eisenmenger Complex
(with right sided aortic arch and descending aorta)

(Age 8 months)

Fig 76 Eisenmenger complex. Notice the large size of the pulmonary artery. The arch of the aorta and the descending portion are to the right of the trachea.

Fig 77 Eisenmenger complex. Same case showing the opened left ventricle and the high ventricular septal defect.

Fig 78 A P roentgenogram of the chest. Notice the cardiac enlargement and the heavy vascular markings of the lungs.

Fig 79 EKG showing right axis deviation, peaked P waves in lead II and diphasic I waves in lead III. The QRS complex is inverted in lead I and upright in lead III.

Fig 80 Eisenmenger complex. Section of aorta and pulmonary artery (longest section). Notice the hypertrophy of the pulmonary artery which is thicker than the aorta. Elastic tissue stain ($\times 10$).

Fig 81 Eisenmenger complex. Section of lung showing the large and thick walled pulmonary arteries. Elastic tissue stain ($\times 10$).



Fig 80

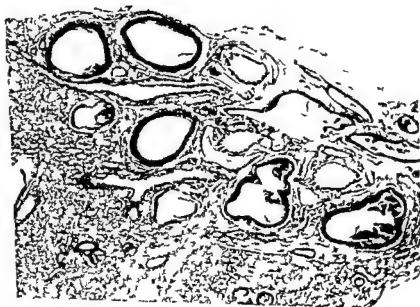


Fig 81

Eisenmenger Complex

(Age 18 months)



Fig 82

Fig 82 Eisenmenger complex. Notice the large pulmonary artery and the small aorta. Notice also the hypertrophy of the right ventricle.

Fig 83 Eisenmenger complex. The probe passes through the ventricular septal defect.

Fig 81 Eisenmenger complex. Sections of aorta and pulmonary artery (longest section). Notice the pronounced relative thickness of the pulmonary artery. Elastic tissue stain ($\times 15$).

Fig 85 Eisenmenger complex. Section of lung showing the large and thick walled pulmonary arteries. Elastic tissue stain ($\times 15$).



Fig 83



Fig 81

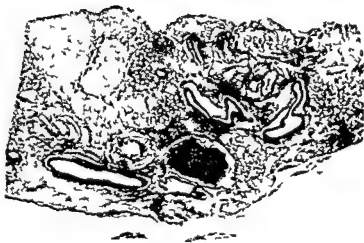


Fig 85



Fig 86

Eisenmenger Complex with Interruption of Aortic Arch

(Age 6 months)

Fig 86 Eisenmenger complex with interruption of the aortic arch. Notice the hypertrophy of the right ventricle and the pulmonary artery. Notice also the interruption of the aortic arch with the pulmonary artery running into the descending aorta through a patent ductus arteriosus. The right subclavian and right carotid arteries originate proximal to the interruption while the left subclavian and left carotid arteries originate in the region of the patent ductus arteriosus.



Fig 87



Fig 88

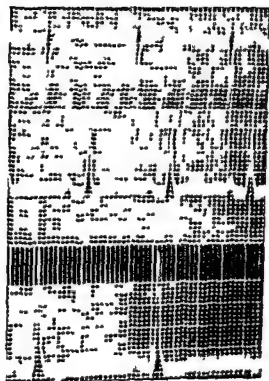


Fig 89



Fig 90

Tetralogy of Fallot (after Potts-Smith operation)

(A 17 year old male)

Fig 87 Heart from a seventeen year old male with tetralogy of Fallot. Notice the large right ventricle, the dextroposed aorta and the large ventricular septal defect. Notice also the rudimentary third (in fundibular) ventricle from which originates a very small pulmonary artery.



Fig 88 A P roentgenogram of the chest. The shape of the heart is normal. The pulmonary conus is slightly concave and the lung fields are avascular.

Fig 89 E K G revealing a very pronounced right axis deviation and peaked P waves in lead II. The Q R S complex is inverted in lead I and upright in leads II and III.

Fig 90 Descending thoracic aorta showing a probe in the surgically created ductus arteriosus. Notice also the large bronchial arteries.

Fig 91 Tetralogy of Fallot. Sections of aorta and pulmonary artery. Notice the very thin pulmonary artery. Plastic tissue stain (X 15).



Fig 92 Tetralogy of Fallot. Section of lung showing the small size and thin wall of the pulmonary arteries.

Fig 92

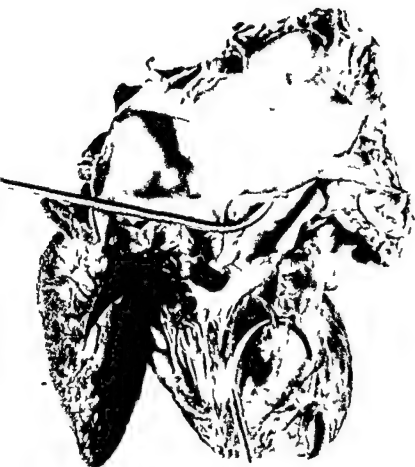


Fig 93

Tetralogy of Fallot

(Age 6 years)

Fig 93 Tetralogy of Fallot Notice the large right ventricle the dextroposed aorta the small pulmonary artery, and the ventricular septal defect

Fig 94 A P roentgenogram of the chest Notice the upturned apex and the avascular lung fields

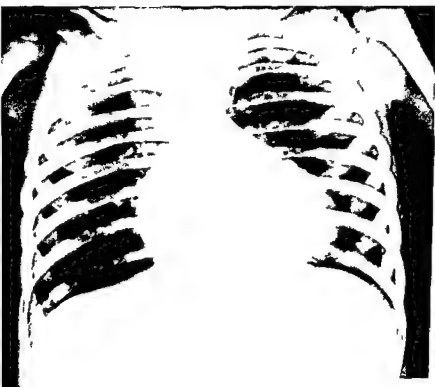


Fig 94



FIG. 93

Tetralogy of Fallot with Bulbar Inversion

(Age 3½ years female)

FIG. 93 Tetralogy of Fallot with bulbar inversion. Notice the hypertrophy of the right ventricle, the dextroposed aorta, the large ventricular septal defect, and the small pulmonary artery behind the aorta.



Fig 96



Fig 97

Tetralogy of Fallot and Dextrocardia (Age 2 years)

Fig 96 Tetralogy of Fallot and dextrocardia. Notice the arch of the aorta to the right of the trachea.

Fig 97 Tetralogy of Fallot with dextrocardia. Notice the anatomical right atrium with a probe in the superior vena cava, and two other probes in the pulmonary veins of the left lung. The other two pulmonary veins drained normally into the anatomical left atrium.

• Taussig-Bing Heart

Pathology (Plate 16)

Aorta arises from the right ventricle
 Pulmonary artery overrides a ventricular septal defect
 Right ventricle and pulmonary artery hypertrophied

Course of the Circulation

Pulmonary artery receives blood from both ventricles
 Aorta receives blood only from the right ventricle
 There is increased flow and pressure in the pulmonary circulation

Clinical Features

Similar to that produced by L₁ emmenget's complex however, in this instance cyanosis is present from birth

Electrocardiogram

Right axis deviation
 Right ventricular preponderance

Radiological Studies

Heart moderately enlarged
 Pulmonary cone is full
 Vascular lung markings increased

Laboratory

Arterial O₂ unsaturation
 Hemoconcentration
 Catheterization reveals O₂ content of the right ventricular blood sample to be increased. Catheter may enter the aorta directly from the right ventricle

Prognosis

Compatible with life for several years

Treatment

None

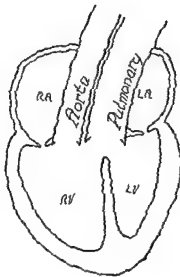


PLATE 16
 Taussig-Bing heart



Fig 96

Tetralogy of Fallot and Dextrocardia

(1yr 2 years)

Fig 96 Tetralogy of Fallot and dextrocardia. Notice the arch of the aorta to the right of the trachea.



Fig 97

Fig 97 Tetralogy of Fallot with dextrocardia. Notice the anatomical right atrium with a probe in the superior vena cava, and two other probes in the pulmonary veins of the left lung. The other two pulmonary veins drained normally into the anatomical left atrium.



Fig 100

Tau sig. Bing Heart

(Age 7 1/2 months)

Fig. 100 AP roentgenogram of the chest taken shortly after birth. Notice the cardiac enlargement especially on the right side.

Fig. 101 AP roentgenogram of the chest of the same patient at 7 1/2 months of age. Notice the enormously dilated globular heart.



Fig 101

Taussig Bing Heart

(Transposition of the aorta and levoposition of the pulmonary artery)

(Age $2\frac{1}{2}$ years)

Fig 98 Heart and lungs from a case of Taussig Bing complex. Notice the large defect in the ventricular septum and the pulmonary valve directly above the defect. Notice also the completely transposed aorta.

Fig 99 A P roentgenogram of the chest. Notice the cardiac enlargement to the right and left, and the increased hilar markings.

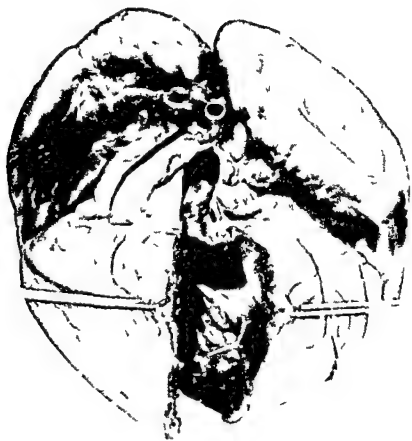


Fig 98





Fig 101



Fig 102

**Corrected Transposition of the Aorta
and Ectoposition of the Pulmonary
Artery**

(Taussig-Bing Heart with Corrected Tran-
sposition of the Aorta)

(Age 14 years)

Fig 102 Heart showing hypertrophy of the right ventricle, the anterior position of the aorta, and the large ventricular septal defect below the pulmonary valve

Fig 103 Heart viewed from the left lateral side. The transposition is readily shown. The ductus arteriosus is patent

Fig 104 Section of lung showing pulmonary arteriosclerosis and necrotizing arteritis. The latter process was confined to the pulmonary arteries





Fig. 104

• Complete Transposition of the Arterial Trunks

Pathology (Plate 17)

- Aorta arises from the right ventricle
- Pulmonary artery arises from the left ventricle
- Aorta situated anterior and to the right of the pulmonary artery
- Ventricular septal defect
- Auricular septal defect
- Patent ductus arteriosus

Course of the Circulation

- Systemic venous blood reaches the right heart and without oxygenation returns to the systemic circulation through the transposed aorta
- Oxygenated blood reaches the left heart through the pulmonary veins and returns to the lungs from the left heart through the transposed pulmonary artery
- Thus there are two entirely separate circulations. This condition is incompatible with extra uterine life unless there are associated abnormalities that permit shunts between the two circulations
- Associated lesions are ventricular septal defects, auricular septal defects, and patent ductus arteriosus. Any one or all of these associated lesions may be present. These compensating shunts are necessary in order to maintain blood volume equilibrium

Clinical Features

- Cyanosis from birth
- Progressive cardiac enlargement begins in early infancy
- Poor growth and development
- Enlarged liver
- Basal systolic murmur

Electrocardiogram

- Right axis deviation
- P waves may be abnormal

Radiological Studies

- Cardiac enlargement
- Shadow cast by the great vessels narrow in AP view
- Shadow cast by the great vessels broad in LAO view
- There is usually evidence of pulmonary congestion
- Angiocardiography demonstrates the transposed aorta and the vessels' anterior position

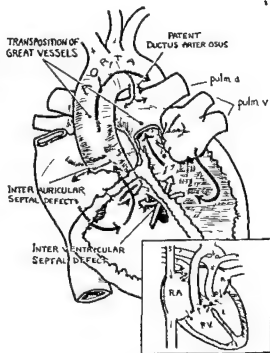


PLATE 17

Complete transposition of the great vessels as associated with ventricular and auricular septal defects and a patent ductus arteriosus

Simultaneous filling of aorta and pulmonary artery will be seen in the angiocardigram if a ventricular septal defect is present

Laboratory

Arterial O₂ unsaturation

Hemoconcentration

Cardiac catheterization may be of value in determining the type and number of associated malformations

Prognosis

Poor—majority of patients with this malformation die in early infancy

Treatment

Experimental Creation of an extra cardiac shunt and an auricular septal defect in our experience, has been of little benefit



Fig 105



Fig 106



Fig 107

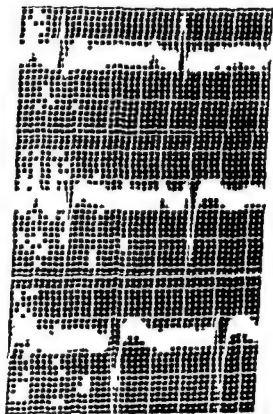


Fig 108

Complete Transposition of Arterial Trunks with "Riding" Tricuspid Valve Type IV of Sputer

(age 8 years)

Fig 10a Complete transposition of arterial trunks Notice the aorta in front and to the right of the pulmonary artery

Fig 10b Same case Posterior view of the heart showing the right atrium and the tricuspid valve Notice how the tricuspid valve extends over a defect in the ventricular septum and opens into both ventricles

Fig 107 A P roentgenogram of the chest Notice the left ventricular hypertrophy the broad supracardiac shadow and the increased pulmonary vascularity

Fig 108 ECG showing a pronounced left axis deviation The QRS complex is upright in lead I and inverted in leads II and III

Fig 109 Complete transposition of the arterial trunks Sections of aorta and pulmonary artery Notice the thick pulmonary artery with a vegetative overgrowth on the intimal surface Elastic tissue stain (X 1a)

Fig 110 Same case Section of lung showing two arteries with an almost complete occlusion of the lumen by concentric layers of fibrous connective tissue Elastic tissue stain (X 200)

Fig 111 Same case Section of lung showing large and thick walled pulmonary arteries Elastic tissue stain (X 1a)



Fig 110



Fig 111



Fig 109



Fig 112



Fig 114

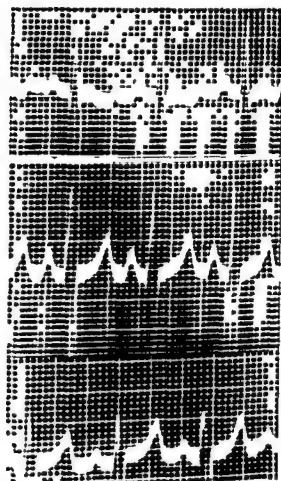


Fig 113

Complete Transposition of Arterial
Trunks with Infundibular
(Subaortic) Stenosis

(Age $3\frac{1}{2}$ years)

Fig. 112 Heart showing the aorta in front
and to the right of the pulmonary artery.
Notice the infundibular (subaortic) stenosis
pre-
sen

Fig. 113 EKG showing moderate left
axis deviation. The QRS complex is
upright in lead I and diphasic in leads II
and III. Peaked P waves are present in
lead II.

Fig. 114 Same case A P roentgenogram of
chest. Notice the cardiac enlargement and
the vascular lung fields.

Fig. 115 Section of aorta and pulmonary
artery. Notice the pronounced thickness of
the pulmonary artery (larger section).
Elastic tissue stain ($\times 15$).

Fig. 116 Section of lung revealing very
prominent and thick pulmonary arteries.
Elastic tissue stain ($\times 15$).



Fig. 115

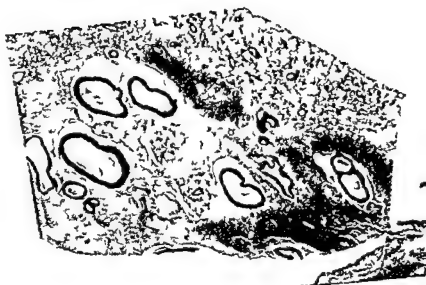


Fig. 116



Fig 117



Complete Transposition of Arterial Trunks

(Age 6 weeks)



Fig 117 Heart and lungs Notice the aorta to the right and slightly anterior to the pulmonary artery

Fig 118 A P roentgenogram of the chest Notice the cardiac enlargement and the narrow supracardiac shadow

Fig 119 Angiocardiograph Notice the simultaneous visualization of the aorta and the pulmonary artery as a result of a ventricular septal defect The aorta originates in the right ventricle

Fig 120 Section of aorta and pulmonary artery Notice the very thick pulmonary artery (longer section) Elastic tissue stain ($\times 10$)

Fig 121 Section of lung showing very large and thick pulmonary arteries Elastic tissue stain ($\times 15$)

Fig 120



Fig 121



Fig 117



Fig 118



Fig 119

Ventricular Inversion

Pathology

Anatomical left ventricle and mitral valve are located beneath the normal right atrium

Anatomical right ventricle and tricuspid valve are located beneath the normal left atrium

Course of the Circulation

Normal

Pulmonary Valve Stenosis with Ventricular Inversion

(Age 1½ years)

Fig 124 Left side of the heart showing the left atrium the tricuspid valve and the anatomical right ventricle. The aortic valve is situated behind the tricuspid valve. The picture also shows the stenotic pulmonary valve with a vegetative process in it.

Fig 125 A P roentgenogram of the chest. Notice the large pulmonary artery and the azygous lumen fields.

Fig 126 Right side of the heart showing the right atrium the mitral valve and the anatomical left ventricle with an added infundibular portion.

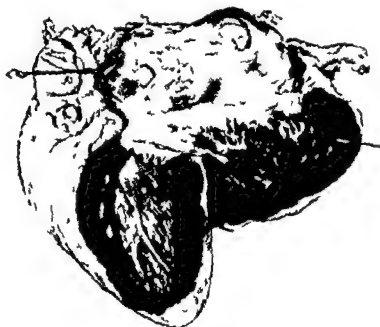


Fig 124



Fig 125

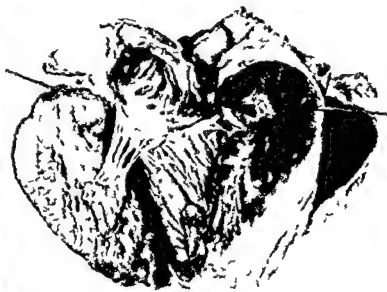


Fig 126

• Corrected Transposition

Pathology

Aorta and aortic valve anterior

Pulmonary artery and pulmonary valve posterior

Each vessel arises from the corresponding ventricle

Course of the Circulation

Normal



Fig 122

Corrected Transposition

(Age 40 years)

Fig 122 Heart from a 40 year old male with corrected transposition of the arterial trunks. The aorta is anterior to the pulmonary artery but originates from the left sided ventricle. Notice the abnormal appearance of the papillary muscles. Notice also the crista supraventricularis running from the base of the aortic valve to the anterior wall of the ventricle.

Fig 123 Section of aorta and pulmonary artery showing normal thickness and relationship.



Fig 123

6

Anomalies of the Aortic Arch

Coarctation of the Aorta

Pathology

- Constriction of the aorta at or near the point of entry of the ductus arteriosus
 - Infantile type — pre ductal
 - Adult type — post ductal
- Pre ductal type usually associated with intracardiac anomalies

Course of the Circulation

- Pre ductal type—right to left shunt through the ductus arteriosus
- Post ductal type—results in development of large collateral channels (internal mammary, intercostal, anastomosis about the clavicle)

Clinical Features

- A common cause of heart failure in cyanotic infant
- Many cases asymptomatic diagnosed on routine examination
- Normal growth and development
- Hypertension in upper extremities
- Weak or absent femoral pulsations
- Heart of normal size
- Soft systolic brachial murmur
- Loud systolic murmur over the left chest posterior

Electrocardiogram

- Left axis deviation
- Left ventricular preponderance

Fig. 127 EKG showing peaked P waves in leads I and II and inverted P wave in lead III. There are also marked right axis deviation and inverted T waves in lead II and III.

Fig. 128 Section of aorta and pulmonary artery. Notice the thin pulmonary wall which reveals a vegetative process at one end.

Fig. 129 Section of lung showing very small and thin walled pulmonary arteries for a 14 year old. Elastic tissue stain ($\times 15$).

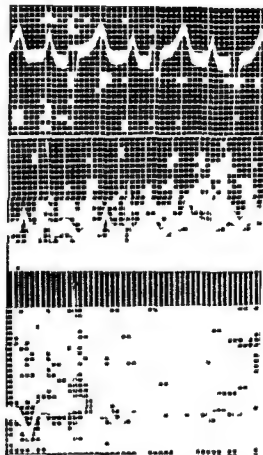


Fig. 127



Fig. 128

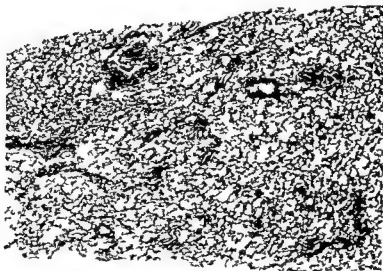


Fig. 129

Coarctation of the Aorta

(Age 21 months)

Fig 130 A P roentgenogram of the chest (pre-operative) from a case of coarctation of the aorta. Notice the marked cardiac enlargement.

Fig 131 A P roentgenogram of same case less than twenty four hours postoperative. Notice the marked decrease in cardiac size.

Fig 132 A line drawing to represent the coarctation and surgical repair in this case.



Fig 131

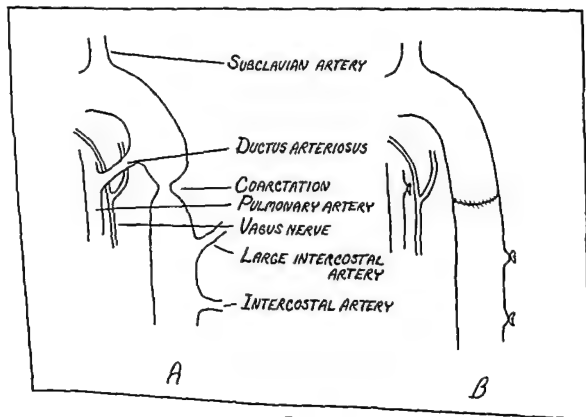


Fig 132

Radiological Studies

Heart of normal size

Notching of inferior rib margin (adults)

Arteriogram reveals constricted area and the collateral vessels

Laboratory

Direct arterial pressure tracings from the brachial and femoral arteries confirm the hypertension in the upper extremities

Prognosis

Good

Treatment

Surgical—resection of constricted area with end to end anastomosis, the use of human arterial graft



Fig 130

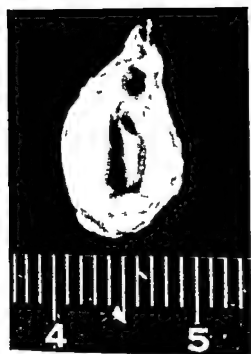


Fig 135



Fig 136



Fig 137



Coarctation of the Aorta

(Age 22 years)

Fig 133 Arteriogram from a case of coarctation of the aorta Notice the constricted area in the thoracic aorta and tortuous internal mammary artery

Fig 134 A line drawing to illustrate the location of the coarctation in this case and the type of surgical removal

Fig 135 Specimen of the same case Notice the marked constriction in the lumen of the aorta

Fig 136 Specimen of coarctation Notice the marked dilatation of the aorta distal to the coarctated area

Figs 137 and 138 A P roentgenograms from two cases of coarctation Notice notching of the undersurface of the ribs as seen in adults

Fig 133

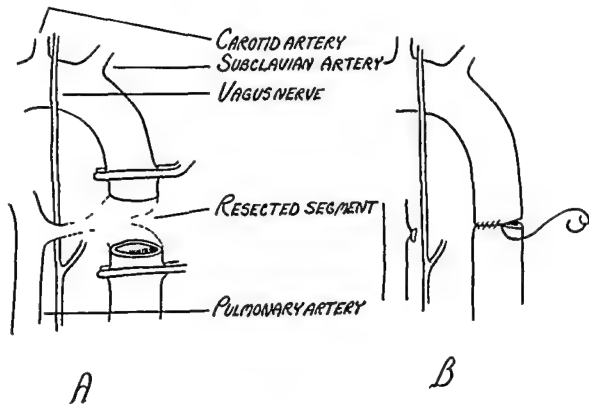


Fig 134



Fig 142

Pre Ductal Coarctation

(Age 1 year)

Fig 142 Heart and great vessels from a one year old child with infantile coarctation. Notice the diffuse narrowing of the arch of the aorta from the point of origin of the left carotid to the point of entrance of the ductus arteriosus.



Fig. 139

Coarctation of the Aorta with rupture of the wall below the point of constriction

(11 year old male)

Fig. 139 Aorta from an 11 year old male with coarctation. Notice the sharply localized constriction, the large upper intercostal arteries, and the insertion of the ligamentum arteriosum.

Fig. 140 A P. roentgenogram from the same case. The heart is normal in size and configuration. Notching of the ribs is not present.

Fig. 141 Specimen from same case that shows rupture of the aorta which occurred below the area of coarctation and into esophagus.



Fig. 140



Fig. 141

Arterial Rings

(Illustrations on page 88)

Pathology

Constricting arterial ring around the trachea and esophagus
Arterial ring usually consists of a double arch or a right aortic arch with a left descending aorta. The ductus or ligamentum arteriosus completes the constricting ring.

Course of the Circulation

There is usually no associated intracardiac anomaly.
The hemodynamics of the circulation are not altered.

Clinical Features

Increased incidence of respiratory infections in early childhood
Dysphagia in early childhood
There may be respiratory distress
No cardiac murmurs

Electrocardiogram

Normal

Radiological Studies

Barium swallow usually reveals a right arch or evidence of a double arch. A retro esophageal vessel is usually discerned.

Prognosis

If the patient survives the first year of life the prognosis is usually excellent. However, stridor, dysphagia and repeated respiratory infections may make surgical intervention necessary.

Treatment

Surgical

Patent Ductus Arteriosus

Pathology (Plate 18)

The ductus arteriosus has its origin at the bifurcation of the pulmonary artery, and its entrance into the aorta is two or three centimeters distal to the point of origin of the left subclavian artery.

Course of the Circulation

The higher pressure in the aorta produces a left to right shunt (aorta to pulmonary) through the ductus.

Clinical Features

More common in the female
Growth and development may be normal or retarded
Heart normal size
Continuous roaring murmur in 2nd left ICS outside the area of cardiac dullness
A marked thrill present in the same area
Peripheral signs of aortic insufficiency

Electrocardiogram

Usually normal

Radiological Studies

Heart usually normal size or slightly large
The left ventricle may be prominent
Increased vascularity of the lung fields
Hilar pulsations noted on fluoroscopy
Appearance of the pulmonary conus not consistent
In atypical cases arteriograms may be of value in diagnosis—
dye can be seen entering the lung fields from the systemic circulation

Laboratory

Catheterization reveals an increase in the oxygen content of the pulmonary artery sample as compared to the right ventricular sample

Prognosis

Good, complications usually do not develop until adult life

Treatment

Surgical closure of the ductus

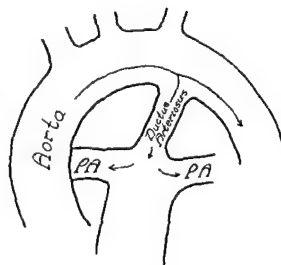


PLATE 18
Patent ductus arteriosus

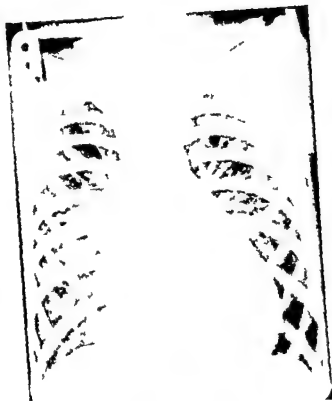


Fig 146



Fig 147



Fig 148

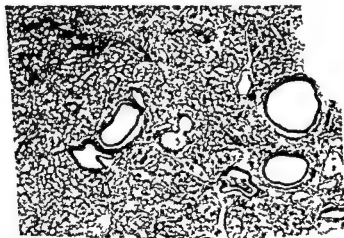


Fig 149



Fig 113

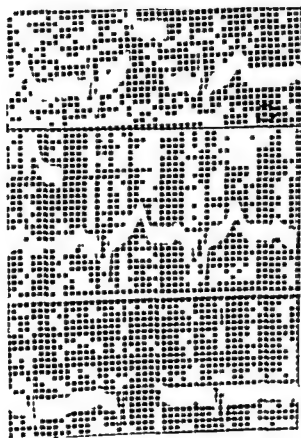


Fig 114

Patent Ductus Arteriosus

(5 year old male)

Fig 113 Heart with a patent ductus arteriosus. The heart is enlarged bilaterally.

Fig 114 The electrocardiogram (standard leads) from the same case reveals a normal tracing.

Figs 115, 116 and 117 AP roentgenograms from children between the ages of seven and nine with a typical patent ductus arteriosus. Notice the difference in the shadow cast by the pulmonary conus in the three cases. Also notice that heavy lung markings are common to all three.

Fig 118 Sections of aorta and pulmonary artery from a case of patent ductus arteriosus. Notice the hypertrophy of the pulmonary arterial wall which is as thick as the aortic wall. Elastic tissue stain ($\times 15$).

Fig 119 Lung section from a case of patent ductus arteriosus showing the large and thick pulmonary arteries. Elastic tissue stain ($\times 15$).

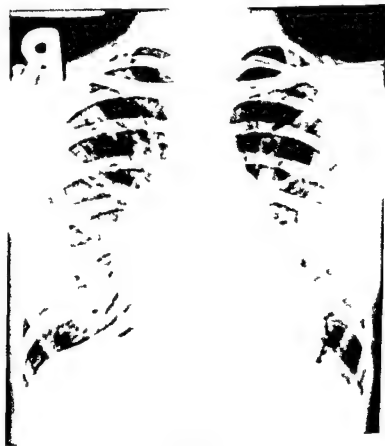


Fig 115

7

Ebstein's Malformation of the Tricuspid Valve

Pathology

This condition consists of a malformation of the tricuspid valve in which there is marked fusion of the leaflets to each other and to the ventricular wall and also a downward displacement of the posterior and lateral aspects of the valve. Moreover, the proximal portion of the right ventricle is incorporated into the huge right auricle and it communicates with the distal (outflow) portion of the ventricle through a large opening in the anterior leaflet of the tricuspid valve. In most cases there is an associated auricular septal defect.

Course of the Circulation

The malformation of the tricuspid valve and the right side of the heart results in an increased pressure and a marked dilatation of the right atrium. Cases with an auricular septal defect develop a marked right to left shunt, with decrease in the pulmonary blood flow and associated cyanosis.

Clinical History

Retardation of growth and development

Delayed onset of cyanosis, disproportion between degrees of cyanosis and dyspnea. Limitation of activity with easy fatigability. No history of squatting. Frequent episodes of paroxysmal tachycardia.

Physical Examination

Mild to severe cyanosis. Cardiac enlargement, mostly right atrium and ventricle. Distant heart sounds, systolic murmur, gallop rhythm.

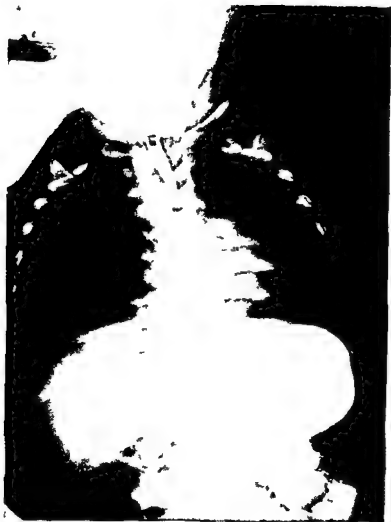


Fig 150



Fig 151

Arterial Ring Around Trachea and Esophagus

Fig 150 A P roentgenogram of chest from a case of vascular ring age 6 months Notice that the barium swallow reveals a bilateral compression of the esophagus

Fig 151 L A O roentgenogram of the chest from the same case Notice the anterior displacement of the barium filled esophagus by the retroesophageal vessel

7

Ebstein's Malformation of the Tricuspid Valve

Pathology

This condition consists of a malformation of the tricuspid valve in which there is marked fusion of the leaflets to each other and to the ventricular wall and also a downward displacement of the posterior and lateral aspects of the valve. Moreover the proximal portion of the right ventricle is incorporated into the huge right auricle and it communicates with the distal (outflow) portion of the ventricle through a large opening in the anterior leaflet of the tricuspid valve. In most cases there is an associated auricular septal defect.

Course of the Circulation

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Clinical History

Retardation of growth and development

Delayed onset of cyanosis, disproportion between degrees of cyanosis and dyspnea. Limitation of activity with easy fatigability. No history of squatting. Frequent episodes of paroxysmal tachycardia.

Physical Examination

Mild to severe cyanosis. Cardiac enlargement, mostly right atrium and ventricle. Distant heart sounds, systolic murmur, gallop rhythm.

Electrocardiogram

Prolonged auricular ventricular conduction Right bundle branch block High and peaked P waves At times, paroxysmal supraventricular tachycardia

Radiological Studies

Clear lung fields Concave pulmonary conus Cardiac enlargement, especially right atrium and right ventricle Angiocardiography reveals pooling of the dye in the enlarged right auricle for too long period of time

Laboratory Studies

Prolonged circulation time Decreased pulmonary blood flow, with right to left flow through auricles Hemoconcentration Arterial oxygen unsaturation

Treatment

Medical No surgical treatment at present

Prognosis

Poor



Fig 152



Fig 153

Ebstein's Malformation of the Tricuspid Valve

(Age 29 years)

Fig 152 Heart and lungs in a case of Ebstein's malformation. Notice the marked dilatation of the right side of the heart and the small size of the pulmonary artery.

Fig. 153 Ebstein's malformation. Anterior view of the right ventricle showing a marked dilatation of the pulmonary conus and a large opening at the base of the anterior leaflet of the tricuspid. The pulmonary valve can also be seen.

Fig 154 Ebstein's malformation. Posterior view of the right ventricle and atrium showing the enormous dilatation of the former. Notice the marked displacement of the posterior leaflet of the tricuspid valve toward the apex of the heart. Notice also the incorporation to the large right atrium of the proximal portion of the right ventricle. In the center of the picture there is the large defect through the anterior leaflet of the tricuspid. On the upper left the patent foramen ovale can be seen.

Fig 155 Ebstein's malformation. EKG showing paroxysmal supraventricular tachycardia, right ventricular preponderance and right bundle branch block.



Fig. 154

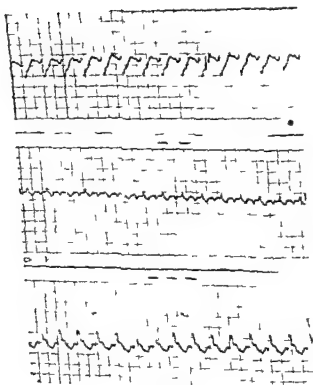


Fig. 155



Fig 156

Fig 156 Ebstein's malformation A P radiograph of the chest showing a marked increase in cardiac size and normal pulmonary vascular markings The pulmonary conus is not prominent



Fig 157

Fig 157 Ebstein's malformation L A O (almost lateral) radiograph of the chest Notice the very large right ventricle which reaches the anterior chest wall



